Papillary Fibroelastoma: A Rare Cardiac Tumor

(A Case Presentation)

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Cardiac papillary fibroelastoma (CPF) is a rare, benign, primary tumor of the heart and accounts for seventy-eight percent of cardiac tumors (Redberg, 1995). The majority of patients with papillary fibroelastomas are asymptomatic although there is a significant risk for cerebral embolization (McFadden & Lacy, 1987).

Reports have associated CPF with coronary, cerebral, pulmonary, and retinal artery emboli (Sun et al.). Papillary fibroelastomas usually arise from the cardiac valves or occasionally from the ventricular endocardium and are most commonly seen in patients over age 50 (Hall, Cooley, McAllister, Frazier, and Wilansky).

Shahian, Labib, and Chang (1995) indicate that studies show a varied age range from the neonate to a 92-year-old patient and that before the availability of the transthoracic and transesophageal echocardiography, papillary fibroelastomas were predominantly discovered on autopsy. The general consensus is that the CPF is being discovered during life with the increased use of echocardiography and transesophageal imaging.

Abstract:

Cardiac papillary fibroelastoma is a rare, benign, primary tumor of the heart and accounts for seventy-eight percent of cardiac tumors. Patient are usually asymptomatic although there is a significant risk for embolization. Papillary fibroelastomas usually arise from the cardiac valves. This is a case presentation of a patient who was admitted to our institution and his course of treatrnent leading to excision of the cardiac papillary fibroelastoma.

The following case describes one patient at our institution who underwent surgical excision of a papillary fibroelastoma.

A.H. is a 45 year old male who initially was seen in the emergency department on May 5, 2001. A.H. had, what he described as chest pain for a "full day". He had been at a party the night before, had a "few drinks" and thought his pain and discomfort were related to this excessive activity. His EKG on admission showed some changes and the attending cardiologist felt these changes might be due to pericarditis. A.H. was admitted to the coronary care unit for overnight observation. After some discussion with the cardiologist, A.H. was discharged the next day with the understanding that he would require an echocardiogram and he would be notified of the date of his appointment. The following day, Monday May 7, 2001, A.H. received a call and was told his appointment was scheduled for July 26, 2001. A.H. felt that this wait was inconsistent with what he had been told by the cardiologist and expressed this fact to the secretary. He still had yet to have a diagnosis and was concerned with this. The secretary called him a short time later and had been able to arrange an appointment for the next day, May 8, 2001.

A.H. returned to the hospital on May 8, 2001 for the echocardiogram (ECHO). The ECHO showed a small growth on the aortic valve. The cardiologists in attendance felt this growth was consistent with endocarditis and A.H. was once again admitted to hospital for a course of intravenous antibiotics.

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Two and a half weeks after the start of treatment, a repeat ECHO showed that there were no changes to the growth on the aortic valve. At this time, the cardiologist elected to perform a transesophageal echocardiogram (TEE). The TEE also showed a small growth which the cardiologist felt was now consistent with findings of papillary fibroelastoma. It was decided at this time that A.H. should be discharged home for one month. He was to "take it easy" and not do anything too strenuous. This was to allow the cardiologist time to confer with colleagues and decide on a further course of treatment. Within the month, the TEE was repeated and again it showed no changes to the tumor. It was decided at this time that surgery would be the next step.

As part of the work-up for surgery, the history and physical for A.H. indicated that he had not experienced any neurological symptoms, with the exception of an occasional headache, no numbness, transient ischemic attacks, or stroke. There was no history of hypertension, diabetes, myocardial infarction, peripheral vascular disease, or congestive heart failure. A.H. had had a tonsillectomy at age 16 years, and previous abdominal surgery in 1984 or 1986. A.H. was a pack a day smoker, who is trying to quit, enjoys a social drink of alcohol, and has a history of increased cholesterol. He is not overweight, does have occasional headaches which he attributes to the stress in his day to day life.

On admission, there was no evidence of carotid bruits, no shortness of breath or cyanosis, the Sl S2 sounds were audible with no S3 S4 or heave or thrill. Prior to surgery, it was felt by the attending physicians, that A.H. should also have a cardiac catheterization, in the event that there was coronary artery disease, coronary artery bypass could be performed at the same time. The cardiac catheterization also ruled out the possibility of valvular damage.

Excision of a CPF

On August 2, 2001, A.H. underwent surgery for excision of a cardiac papillary fibroelastoma. Open heart surgery was performed through a median sternotomy. Cardiopulmonary bypass was instituted and an incision was made in the aortic root above the cusp of the aortic valve. The papillary fibroelastoma was identified on the ventricular surface of the left coronary cusp of the valve and subsequently excised. The aortotomy was repaired, the patient stabilized, and cardiopulmonary bypass discontinued. It was during this time that the patient experienced ventricular fibrillation, was unstable, and cardiopulmonary bypass was reintroduced. Fibrillation occurred during protamine administration and the patient was extremely unstable during this time.

Once again, the patient was stabilized and cardiopulmonary bypass was successfully discontinued. It was discussed between the surgeon and the anesthetist and they agree that the patient may have had an allergic type reaction to the protamine. This could potentially explain the patient sudden response, fibrillation and instability for an otherwise stable situation. The patient was then transferred to the cardiac intensive care where he had an uneventful postoperative recovery. A.H. was discharged home on August 6, 2001 to return for follow-up in six weeks.

Conclusion

Literature is scarce when searching for information on the cardiac papillary fibroelastoma. One thing that all evidence points to, is that the CPF is a rare tumor with the majority studied after autopsy and that only recently with the advent of transthoracic and transesophageal echocardiography, is diagnosis during life possible. Patients with CPF are usually asymptomatic however, neurological sequelae is a common presentation. As demonstrated with this case presentation, the use of the echocardiography and the TEE led to the detection, diagnosis, and subsequent treatment of the tumor. Sun et al (2001) certainly indicate that there are many CPF that go undetected with ECHO. The reasons they give are:

1) the tumor was masked by an associated lesion;
2) the tumor was too small to be seen;
3) the examination was not done carefully with a sufficient amount of suspicion; or
4) there were no significant characteristics to differentiate the CPF from the degenerative valve disease.

This could very well have been one of the reasons for this case to initially to be diagnosed as endocarditis and also indicates the rarity of the